Laurence-Moon Syndrome

Alternative Names
Laurence-Moon-Biedl-Bardet syndrome
Laurence-Moon-Biedl syndrome

WHO International Classification of Diseases
Congenital malformations, deformations and chromosomal abnormalities

OMIM Number
245800

Mode of Inheritance
Autosomal recessive

Description
Laurence Moon syndrome is a rare disorder characterized by hypogonadism, retinitis pigmentosa, mental retardation, and spastic paraplegia. This disorder was earlier treated as a part of Bardet-Biedl syndrome (BBS). However, it is now considered a separate entity from BBS by many, and can be differentiated from it based on the presence of spastic paraplegia, and absence of the obesity and polydactyly, characteristic of BBS. The major symptoms of Laurence Moon syndrome, mentioned above, present during childhood. Other complications of the disease include diabetes mellitus, renal abnormalities, psychiatric illness, developmental delay, abnormal facies, and congenital heart disease, among others.

World over, the disease is rare, affecting only 1 in 160,000 people. However, it shows a higher rate of prevalence in the Arab population. Diagnosis depends entirely on the clinical features. Laurence Moon syndrome is incurable, and therefore, persists as a chronic condition. However, timely symptomatic treatment ensures a good prognosis.

Molecular Genetics
Laurence Moon syndrome is transmitted in an autosomal recessive fashion. Linkage studies have shown the disease to be linked to the chromosome 11q region, just like BBS. This has prompted recent researchers to suggest that BBS and Laurence Moon syndrome are the same entity, with variable expression of symptoms.

Epidemiology in the Arab World

Kuwait
Farag and Teebi (1988) indicated that in the Arab population of Kuwait, 26 cases in 15 families were ascertained to have Bardet-Biedl syndrome (20 cases in 13 families) or Laurence-Moon syndrome (6 cases in 2 families).

United Arab Emirates
Abou-Chaaban et al. (1997) studied the pattern of pediatric renal diseases among children in the Dubai Emirate during the period from 1991 to 1996. In this period, a total of 712 pediatric patients, including 230 nationals of the United Arab Emirates, were seen with various renal problems. Among 50 children with chronic renal failure, Abou-Chaaban et al. (1997) observed 2 individuals with Laurence-Moon-Biedl syndrome.

Yemen
Levy et al. (1970) reported three cases of Laurence-Moon-Biedl-Bardet syndrome in a Jewish Yemenite family. No further details could be obtained as of date.

References

Contributors
Ghazi O. Tadmouri: 29.8.2006
Ghazi O. Tadmouri: 27.8.2006
Pratibha Nair: 27.8.2006